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Case report: Successful treatment of recurrent chordoma and bilateral pulmonary metastases following an 11-year disease-free period

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ABSTRACT

INTRODUCTION: Chordomas are rare but aggressive tumors due to local recurrence and distant metastases. They originate commonly in the sphenooccipital and sacrococcygeal regions, and metastasize to the lungs, bone, skin, liver, and lymph nodes. They occur more frequently in men and people over the age of 40.
PRESENTATION OF CASE: A 28 year-old female presented with sacrococcygeal chordoma for which she received wide local excision and adjuvant radiation therapy. She enjoyed an unusual disease-free survival for 11 years until a routine surveillance scan of the pelvis identified local recurrence. Further work up revealed bilateral pulmonary metastases. She underwent local excision of the recurrent tumor and video-assisted thoracoscopic (VATS) wedge resection of pulmonary metastases. She also received adjuvant radiation therapy to the recurrent resection bed. Two years later, she remains free of disease and symptoms.
DISCUSSION: Chordomas are commonly insensitive to chemotherapy and radiation, making surgery the most successful therapeutic modality. However, there are few guidelines on the surveillance and treatment of recurrent chordoma. We report success with aggressive surgical resection of recurrence and metastasis as well as adjuvant radiation therapy.
CONCLUSION: The prolonged survival of our patient underscores the importance of (1) aggressive surgical resection of chordoma, whether primary, recurrent, or metastatic, with adjuvant radiation therapy, (2) minimization of surgical seeding of tumor, and (3) diligent cancer surveillance.
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1. Introduction

Chordomas are locally invasive, aggressive tumors of notochordal remnants. Chordomas have a general incidence of 0.08 per 100,000 people and constitute 1–4% of all bone cancers.[1] They are especially rare in people younger than 40 and more frequent in men than women.[2] The median survival is 6.29 years with a 10 year survival of 39.9%.[2] Most primary tumors originate in the cranium (32.0%), spine (32.8%), or sacrum (29.2%).[2] Extra-axial metastasis occurs in 3–48% of patients.[3–5] Complete surgical resection is the main modality of treatment. Chordomas are frequently insensitive to chemotherapy and radiation therapy. Local recurrence is the most important indicator of poor prognosis.[6] Metastasis, commonly occurring in the lungs, liver, bone, lymph nodes, and skin, is often associated with local recurrence of the primary tumor.[5] Here, we describe the clinical course and examine the outcomes of our management of a 39-year-old woman who presented with multifocal metastases to the lungs and local recurrence in the gluteal region 11 years after resection of a sacral chordoma.

2. Presentation of case

2.1. Primary treatment: aggressive resection with adjuvant radiation

A 28-year-old woman presented with right buttock discomfort increasing over five years. Physical examination revealed a firm, immobile mass posterior to the rectum. MRI demonstrated a 20 cm mass originating from the sacrum at the level of S5 and extending into the gluteus muscles. A CT-guided biopsy of the mass demonstrated chordoma. The patient underwent *en bloc* resection of the tumor and right vertical rectus abdominus myocutaneous flap for perineal reconstruction. A 6-week course of adjuvant external beam radiation therapy (6000 cGy) was administered. Apart from occasional fecal incontinence, the patient remained otherwise asymptomatic for 11 years with no evidence of local recurrence or metastasis. During this period, she underwent yearly surveillance abdomen and pelvis contrast enhanced CT scans.

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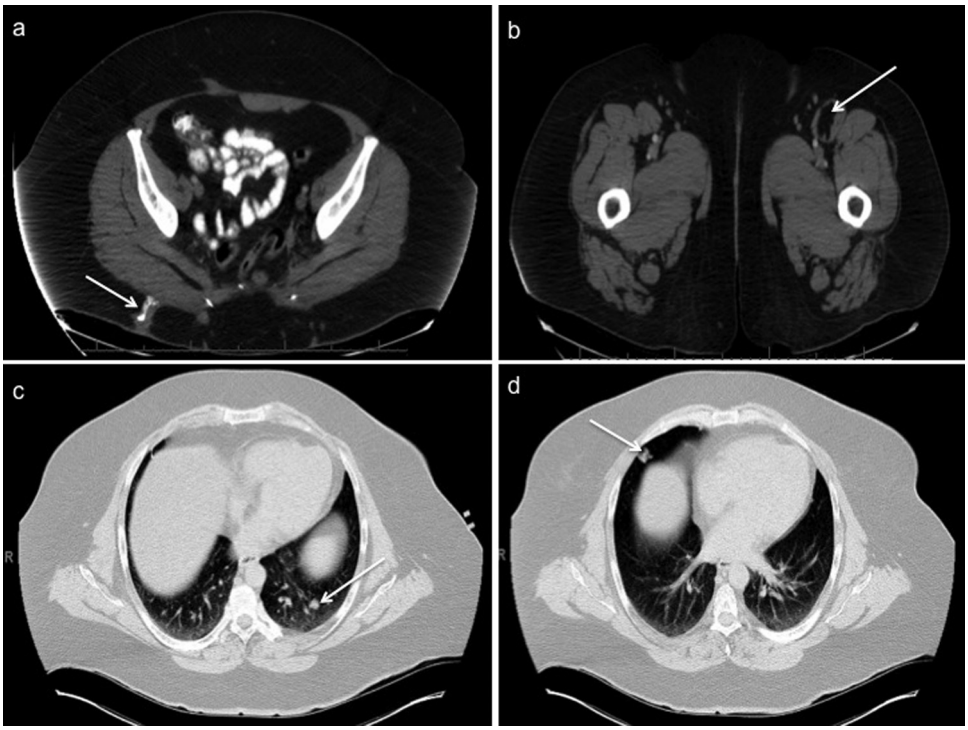

Fig. 1. Axial CT slices with arrows indicating (a) local recurrence of chordoma posterior to right gluteus maximus, (b) mass eventually identified as a lipoma in the left sartorius muscle, (c) metastatic chordoma nodule in left lower lobe of the lung, and (d) metastatic chordoma nodule in right middle lobe of the lung.

2.2. Wide excision of local recurrence and metastasis with curative intent

Eleven years after her initial diagnosis, surveillance CT of the abdomen and pelvis identified a calcified soft tissue density extending into the right gluteus muscle and a 19 mm fatty density in the proximal left sartorius muscle (Fig. 1a and b). The abdomen CT also revealed pulmonary nodules (Fig. 1c and d). A dedicated CT scan of the chest subsequently identified four discrete pulmonary lesions: a 19 mm nodule in the right middle lobe, 3 mm nodule in the superior segment of the right lower lobe, 10 mm nodule in the left lower lobe, and a 10 mm nodule in the left upper lobe. Only the right middle lobe lesion was hypermetabolic by PET scan. Head MRI showed no evidence of intracranial metastasis. CT guided biopsy of the right middle lobe nodule showed metastatic chordoma consistent with the original sacral chordoma. We did not biopsy the suspected site of local recurrence.

Given the appearance of locally recurrent disease with oligometastatic disease, we endeavored to resect all suspected foci of disease with curative intent. In an effort to minimize the number of operations due to the patient's limited financial means while avoiding bilateral lung surgery at the same setting, we planned two surgeries. The patient first underwent video assisted thoracoscopic surgery (VATS) removal and diagnostic biopsy of the two left-sided pulmonary nodules. Both left lung nodules were metastatic chordoma resected with negative margins. A month later, the patient had wide local excision of right buttock mass, right sartorius mass, and VATS resection of two right-sided pulmonary nodules. All lung specimens were removed via a retrieval bag (ENDOPOUCH Specimen Retrieval Bag System, Ethicon, Cincinnati, OH). The right buttock mass was dedifferentiated chordoma with a high-grade spindle cell sarcomatous element and negative margins (Fig. 2). The sartorius mass was an intramuscular lipoma. The right middle lobe lesion was chordoma with negative margins; the right lower lobe lesion was an intrapulmonary lymph node. The final diagnosis was local recurrence and dedifferentiation of sacral chordoma with bilateral pulmonary metastases demonstrating original chordoma histology. After surgery, the patient received 4600 cGy of radiation in 200 cGy doses to the region of the recurrent right buttock chordoma.

2.3. Long-term follow-up

She is free of disease and symptoms two years after her recurrence with metastases and 13 years after her initial diagnosis of chordoma. She has continued to undergo surveillance contrast enhanced CT scans of her chest, abdomen, and pelvis every six months.

3. Discussion

3.1. Pathological change in local recurrence

Dedifferentiation of chordoma has been reported rarely in local recurrence, and has been associated with poor prognosis.[7–9] It is unclear if sarcomatous transformation is due to (1) spontaneous change of the original chordoma, (2) irradiation-induced change, or (3) polyclonal convergence of two different cancers.[9] Our patient is the first known report of dedifferentiated chordoma recurrence with differentiated chordoma pulmonary metastases. The lung metastases could have occurred early, before transformation of the initial chordoma and taken an occult, insidious course, or they could have evolved from differentiated chordoma cells in a polyclonal tumor recurrence. The literature supports the latter scenario as local recurrence typically precedes metastasis.[18]

3.2. Surgery

Aggressive *en bloc* surgical treatment has been associated with decreased local recurrence rates, decreased incidence of metastasis, and overall improved survival.[3,10] Since chordomas tend to be locally destructive tumors, *en bloc* resection may require tissue

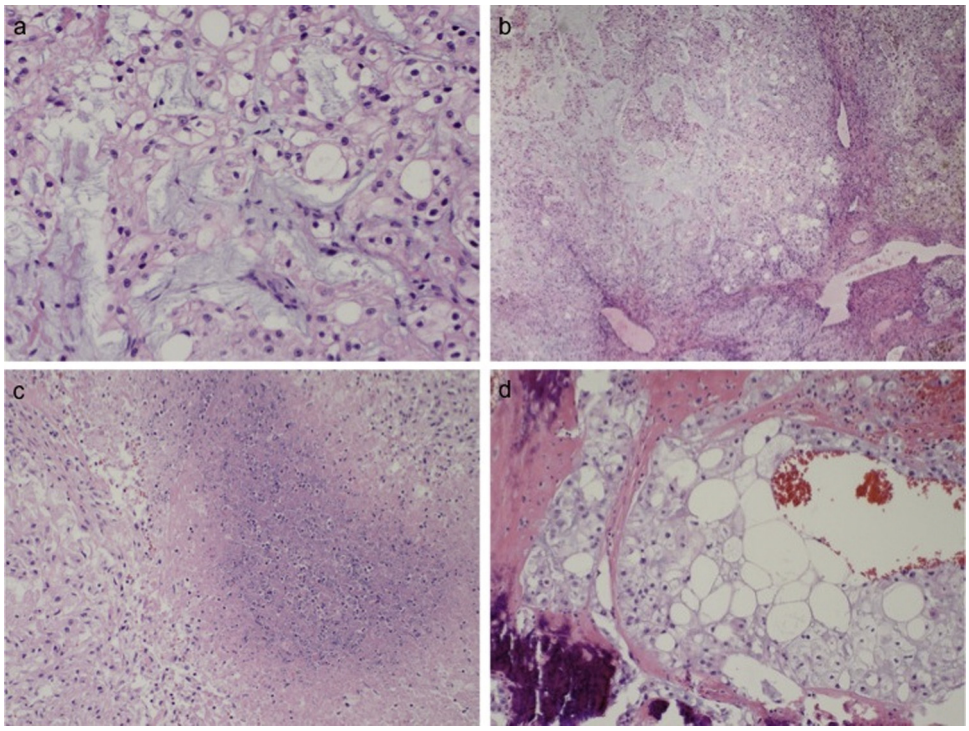

Fig. 2. Pathology H&E stains of gluteal mass showing (a) chordoma cells with characteristic physaliferous cytoplasm and myxoid stroma (20×), (b) lobulated growth pattern of the tumor (4×), (c) zonal necrosis in the area of the tumor (4×), and (d) tumor invading the bone (4×).

reconstruction.[10,11] In our patient, *en bloc* resection of the sacral tumor with negative tissue margins and reconstruction of her perineum led to a long disease free survival after initial treatment. We therefore advocate aggressive initial surgical resection.

Local recurrence can originate from seeding of the tumor during biopsy, resection of the primary tumor, or reconstruction.[12,13] To minimize the local recurrence, we recommend (1) wide resection margins, (2) minimal handling of the tumor, and (3) new instruments, and gloves be used for reconstruction and closure.[13] With pulmonary metastasis, we advocate a VATS wedge resection with wide margins and specimen removal via a retrieval bag to prevent tumor contamination of port sites.

3.3. Radiation therapy

The use of radiotherapy to treat primary and secondary chordoma is controversial, mitigated by the intolerance of adjacent spinal cord and brain stem to high radiation doses.[14] High dose radiotherapy with surgery has been proven to favorably affect disease free interval.[15] Conventional radiation therapy at 40–60 Gy has resulted in 5-year local control of 10–40%.[14,16] Our patient was treated with adjuvant radiation therapy for six months following initial resection and also received adjuvant radiation to the site of local recurrence. While the efficacy of adjuvant radiation therapy in the setting of recurrence is unknown,[17] adjuvant radiation therapy may be a contributor to her continued long-term survival.

3.4. Follow-up

There are poor evidence-based recommendations on chordoma patient follow-up. After initial treatment, patients are followed with frequent office visits in the immediate post-operative period and then yearly surveillance abdomen and pelvis CT scans. Clinical suspicion for local recurrence should be higher in patients who underwent incomplete excision of their original chordoma. Any new nodules, fistulas, draining sinuses, or pain and discomfort in an area even remote from the previous excision warrant investigation for possible recurrence. Local recurrence typically precedes metastasis.[18] Therefore, we recommend that patients with local recurrence should undergo an evaluation for metastasis with full body PET/CT scan and head MRI. The role of routine surveillance for distant metastasis is unknown at this time.

3.5. Treatment of recurrent and metastatic disease

Even with recurrence and metastasis, long-term survival can be achieved with successful surgical resection of all appreciable disease, as seen with our patient. Aggressive *en bloc* resection of the recurrent tumor with seeding precautions is again preferred. There are no reports, let alone consensus, about the treatment of pulmonary metastases in chordoma. We therefore extrapolated our experience with other pulmonary metastasis, electing for VATS wedge resections of each of the pulmonary nodules with negative margins.[19,20] For metastatic chordoma to the lung, we recommend VATS resection if (1) the primary or recurrent tumor can be completely resected, (2) there are no other metastases, and (3) the lung metastases can be resected with ample margins without compromising the patient's quality of life. Even multiple, bilateral pulmonary metastases can be aggressively treated with long-term success.

4. Conclusion

The treatment of chordoma demands both aggressive resection and cautious attention to minimizing surgical tumor implantation. Adjuvant therapy is controversial but known to contribute to long-term disease control and survival. Postoperative patient follow-up should continue long-term; signs or symptoms consistent with possible local recurrence or metastasis should prompt a thorough work-up. In the absence of symptoms, surveillance CT scans can detect potential treatable recurrence. Recurrent disease

can be treated aggressively with curative intent including surgery and adjuvant radiation therapy.

Conflict of interest

The authors have no conflicts of interest to disclose.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request

Author contributions

CPE: Study concept, data collection, data analysis, writing paper. RJB: Study concept, data collection, reviewing manuscript. VR: Study concept, data analysis, writing paper.

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